Scripta Scientifica Medica, vol.28, Suppl.1,pp.173-174

RELATIVE PART OF INBORN DISEASES AS A CAUSE OF DEATH AMONG CHILDREN UP TO ONE YEAR OLD

I. Krasnaliev, M. Gardevski
Department of Pathology, Varna

The decrease of children's mortality is still a priority task of our health care institutions. Many previous investigations show an increasing frequency of inborn malformations in recent decades. WHO reports for about 40% structural abnormalities in new-born. Steady trends of increasing mortality rates of malformations are evident from other studies.

We studied the causes of death of children aged up to 1 year obducted for the period 1988-1989 in the Department of Pathology, Medical University, Varna. All necessary data were provided from clinical and autopsy records.

A total of 256 children (139 male and 117 female) were obducted in that period. Most of the children were full-termed and in the age group from 1 month to 1 year. The inborn abnormalities were the most frequent cause of death - 94 (36,72%), followed by immaturity of organs and systems -62 (24,22%), inflammatory diseases - 48 (18,75%), hyaline membrane disease - 29 (11,33%) and others 23 (8,98%). From all 256 cases we found 111 (43,46%) inborn diseases and malformations. Most frequent were multiple malformations of 2 or more systems - 28 (25,42%), next coming abnormalities of cardiovascular system - 16 (14,55%), nervous system -14 (12,73%), inborn infections - 13 (11,82%), malformations of alimentary tract - 12 (10,91%), mucoviscidosis -7 (6,35%), malformations of urinary system - 6 (5,45%), morbus Down -6 (5,45%), diaphragmatic hernia - 3 (2,73%) and others (systematic skeleton errors, Di Giorgi syndrome, mesenteric cyst, inborn struma diffusa, liver cirrhosis and ganglioneuroma) - 6 (5,45%).

The increased percentage of multiple malformations is evident. This data differs from that in the literature. Zlateva & Kulova report 38,70% multiple malformations, Agopyan - 13,63%, Raichev - 3,14%. The analysis of this group showed that in 9 cases 2 systems were damaged, in 9 - 3 systems, in 9 - 4 systems and in 1 case - 5 systems.

Cardio-vascular system was most frequently involved in multiple malformations - 19 cases, followed by alimentary tract - 12, facial abnormalities - 11 and others. In only 1 case genetic analysis showed trisomia 18 (Edward's syndrome). No other data from genetic

councelling were obtained. In our opinion, most cases should be concerned as unclassified complexes of inborn abnormalities. Among the group of malformations of cardio-vascular system 5 children had endocardial fibroelastosis, 5 - morbus Roger and 6 had combined cardiac defects. In the group with malformations of nervous system 9 children had hydrocephalia with unknown etiology, 2 porencephalia and one - spina bifida, Tay-Sachs disease (amaurotic familial idioty) and angiomatosis of meninges each. Oesophageal atresia was the most frequently isolated malformation in the group with abnormalities of alimentary tract - 6 cases, followed by Ledd syndrome - 3 cases, atresia of duodenum, atresia of colon ascendens and inborn megacolon. All children in this group died from inflammatory complications after operative corrections. From all 13 cases with inborn infections 3 children were with cytomegalovirus infection, 2 with toxoplasmosis, 2 with meningoencephalitis with unknown origin and one - with inborn hepatitis, lues, sepsis and pyoderma each. Generalized form of mucoviscidosis was found in 2 children; in 3 pancreas and intestines and in 2 lungs and intestines were involved. Among the malformations of urinary system most frequent were polycystic kidneys. Inborn abnormalities were cause of death in 94 (85,45%) from all 111 cases (alone or by their complications). Other 16 cases (14,55%) were not straightly involved in tanatogenesis, i.e. they were background diseases. In 43 cases (39,09%) in clinical records data were found for risk factors for delivery new-born with malformation - viral infection in the first trimester of pregnancy, EPH gestoses, hydramnion, multiple pregnancy, previous spontaneous abortions, previously born children with malformations.

In conclusion, from our data is evident that malformations and inborn diseases are the greatest relative part among the reasons for children's death up to 1 year's age.

Our studies correspond with such ones of other authors that show lasting tendency of increasing mortality rates of these diseases. That may be due to increasing the population frequency of inborn diseases and malformations or to decreasing the mortality rates of other diseases in early childhood. Early establishing and elimination of teratogenic factors and early antenatal diagnosis are untapped resources for decreasing children's mortality.